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MITRAL STENOSIS

Mitral stenosis is that anatomical deformity of the mitral curtains whereby the mitral orifice is narrowed. It is of chief interest because it is the most characteristic lesion of rheumatic heart disease. It has been said of rheumatism that "it licks the joints and bites the heart." It may fatally injure the heart in the absence of all joint pain, or it may cause great pain in the joints and leave the heart uninjured.

The usual steps in the formation of the final rheumatic lesion are inflammation, with thickening, deformity, welding, or a contracting scar, resulting in stenosis. Obstruction of the mitral valve may be caused also by a ball thrombus or tumor in the left auricle, aneurysm of a mitral curtain (rare), a bulging ventricular septum from extreme right ventricular hypertrophy, vegetations of bacterial endocarditis, rare congenital deformities, a tumor of the left ventricle, a calcified mass behind a mitral curtain, or a torn valve curtain. Such instances of obstruction may produce a diastolic murmur which imitates the murmur of mitral stenosis, but the characteristic curtain deformity with welding and mechanical obstruction originates practically always with rheumatic endocarditis.

History: Vieussens, in 1715, described a case of mitral stenosis discovered at autopsy, and stated that "the mouth of the left ventricle appears very small" and that it had "an oval, oblong shape." Morgagni described a case in a man 62 years of age. Corvisart, in 1806, referred to " a certain thrill (bruissement) difficult to describe, perceptible when the hand is applied to the precordial region." Laennec described the purring thrill and the blowing murmur. Bertin, in 1824, wrote, — "the only thing truly important is to know whether any orifice be contracted." James Hope mentioned "the associated murmur from regurgitation, even when the diastolic murmur is feeble, soft or absent." A. Fauvell, in 1843, referred to the "intense, rasping murmur (bruit de rape), preceding the first sound, finishing with it." C. J. B. Williams, who invented the two-eared stethoscope, in 1835 noted the timing of the murmur with the "diastole of the ventricle." In 1836, Bouillaud showed that in acute rheumatic fever inflammation of the heart usually occurs, that the valves are damaged, and that this damage is usually permanent. Modern diagnosis has been aided by the use of the Roentgen ray, of the sound-record of the diastolic murmur in its relation to the first and second heart sounds, and of the electrocardiogram. Carey Coombs, in Bristol, England, stressed the importance of rheumatic fever as the most common etiologic factor.

Etiology: Mitral stenosis may be congenital or atherosclerotic, as well as rheumatic, in origin. Abbott has collected six cases of the congenital type. The atherosclerotic type of mitral lesion is relatively rare, and when it occurs is usually associated with a similar change of the aortic curtains. At autopsy the mitral ring shows whitish areas of calcification. The fibrous tissue is increased; the curtains are deformed; and the openings are narrowed in varying degrees. The patients with this type of mitral stenosis are usually past 50 pears of age. Widespread arteriosclerosis, nephritis and hypertension are often present. Diastolic murmurs may be either absent or too low-pitched to be heard, but apical systolic murmurs are usually present and high-pitched. Teleoroentgenograms may show spots of calcification or crescents in the arch or knob of the aorta, and sometimes in the mitral ring or valve, suggesting changes in the aortic and mitral curtains.

Rheumatic Mitral Stenosis

The incidence of rheumatic mitral stenosis corresponds geographically with that of rheumatic fever, which is a disease occurring more frequently in regions having cold climates, long winters, little sunshine, and damp weather. Rheumatic fever appears often to be associated with poverty, crowded quarters and exposure to the elements. It is of common occurrence in the British Isles and in Northern Europe, and is less frequently found in Egypt and Italy. The general practitioner in the Southern States encounters, on an average, one or two cases annually; in the Tropics the disease is even more rare. Among 5606 children who entered the Egleston Hospital in Atlanta, only 12 cases of acute rheumatic fever are recorded, 40 cases of rheumatic heart disease and 38 cases of chorea. The increase in morbidity from heart disease in northern countries has been ascribed to the prevalence of rheumatic fever in colder regions. Mitral stenosis is likewise rarely found in southern hospitals, and is frequently seen in northern hospitals. My own records show 69 cases of mitral stenosis in 9908 private cases, or 0.7 per cent. This disease is rare in the southern Negro, in whom syphilitic aortic disease is frequently found.

Females are more often affected than males, the ratio of incidence being 3:2 or even 2:1. Coombs found this disease in an average of two females to one male, while my own smaller series shows an even greater proportion of women. The fully developed lesion rarely occurs before 10 years of age, but cases have been seen in patients as young as 4 years of age.

Pathology: The term "rheumatic mitral disease" includes both mitral incompetency and mitral stenosis. Either of these conditions may predominate over the other; and, rarely, one is present in the absence of the other. Rheumatic damage results from an inflammation which is proliferative and exudative in character. Endothelial cells may undergo coagulative necrosis, become covered with fibrin, and granulocytes appear in the affected region. In early inflammations sessile vegetations often appear on the auricular surface at the edge of the mitral curtains forming a ruffle-like growth. Beneath the endothelium and within the valve substance the inflammation is widespread. The involved tissue is characterized by the appearance of new vessels, fibroblastic proliferation, and the eventual formation of miliary nodules. As the inflammation recedes, the thin, pliable edge of the normal valve curtain becomes thicker, rolled and shortened, and fails to meet its fellow in systole, causing regurgitation. Repeated attacks with renewed inflammatory changes cause welding of the curtains at their lateral margins and increasing valvular fibrosis; as the chordae tendinae likewise are involved, they fuse, mat and shorten, pulling down the curtains toward the papillary muscles. In time the contraction and increasing rigidity of the curtain may change the normal form of the mitral valve into that of a funnel pointing downward into the ventricle with a mere cleft for

an opening. The circumference of the normal mitral valve is from 9 to 11 cm., measuring rather less in the female than in the male. The stenotic mitral valve generally has a circumference of less than 6½ cm.

Functional Alterations of the Heart: In mitral stenosis, the burden of overwork falls upon the left auricle, the pulmonary vessels and the right ventricle. Ultimately, if the tricuspid ring dilates, the right auricle also is affected, and a rising pressure within the great veins results, which soon causes enlargement of the liver, hydrothorax, ascites and dependent edema.

Symptoms: There is no single symptom that is characteristic of mitral stenosis. As a rule, except in extreme cases, the younger the patient the less he tires on effort and the fewer his complaints; the older the patient, the more he complains of easy fatigability particularly on effort. Many women discover their dyspnea while at housework, and many men while at manual labor. The earliest symptoms referable to the heart are dyspnea, palpitation, tachycardia, cough, dizziness and edema. The patient's family may notice the blue tinge of cyanosis in the patient, or the malar flush and contrasting pallor. Hemoptysis or pedal edema may bring the patient to the physician for examination. Physical examination of students, nurses or insurance applicants may disclose signs of an unknown mitral lesion, with no history of any previous limitation of activity.

Physical Signs: When all the signs are present, the diagnosis is relatively easy to make; however, it is often missed even by careful examiners when there are missing links in the chain of evidence.

On inspection, the patient may present a malar flush, and generally shows cyanosis. Other patients may evince "mitral dwarfism" or limitation of speech and movements.

On palpation, pulsations in the second and third left intercostal spaces may occur. A strong valve closure ("shock") at the apex, and another at the pulmonary area generally are noted. A diastolic thrill is commonly felt in thin persons from midprecordium to apex, and in plump persons is perhaps barely felt at the apex.

The most characteristic auscultatory sign is a low-pitched, rough, rumbling, grating or rasping, mid-diastolic and presystolic murmur, usually crescendo and always ending abruptly with the loudly accentuated first sound of the heart. It is best heard immediately after exercise with the patient lying on his back or on his left side; adminstration of amylnitrite or nitroglycerine may rarely be necessary to make it audible. In auricular fibrillation the presystolic murmur may disappear or become fainter, and the early diastolic element more evident. The murmur is best heard at the apex, or within and above. The sound of the pulmonic valve closure is accentuated and often split, since the pressure in the pulmonary circuit is increased. The systolic murmur of mitral insufficiency usually follows the accentuated first sound, and is often limited to a very small area. It is generally widely transmitted over the precordium and to the left thorax posteriorly. A mid-diastolic murmur occurring in an acute rheumatic attack usually means dilation of the left ventricle but often it is the earliest diastolic sign of mitral stenosis, antedating the presystolic roll and thrill. The opening snap of the mitral valve, an early third sound, is a common sign. It may require two years or longer for the typical physical signs of an advanced mitral stenosis to develop.

The pulse-volume is generally small; and the pulse-rate may be slow in the absence of heart failure, but is often increased with rapid acceleration on exercise. In men the rates commonly range from 80 to 90, and in women from 90 to 100. Uncomplicated cases show an average blood pressure of 100 to 120 systolic over 70 to 90 diastolic, but hypertension may be associated with this condition.

In adults, unless the stenosis is slight or moderate, the vital capacity is usually decreased.

Roentgen-Ray and Electrocardiographic Findings: The heart shadow seen on fluoroscopic examination is characteristic when mitral stenosis is present. The left border is less concave and straighter than normal. The aortic knob appears relatively less prominent. These changes are due to dilation of the pulmonary conus. The left auricle hypertrophies and dilates and may rarely reach a capacity of 600 c.c. or more. Passing back of the heart and to the right it may push the esophagus backward and to the right, and may even appear to the right of the heart. The left recurrent laryngeal nerve may be compressed by such tremendous auricular enlargement, causing hoarseness by paralysis of the left vocal cord.

The electrocardiogram usually shows a right axis deviation, probably due to the preponderant hypertrophy of the right ventricle, but if aortic insufficiency is likewise present, left axis deviation may be the result. Notching and prolongation of the P-wave is usually referable to the left auricular enlargement. Instead of notching, the P-wave may have a broad plateau.

Complications: About 5 per cent of patients develop subacute bacterial endocarditis. More than 80 per cent die of congestive heart failure. Auricular fibrillation occurs in probably more than 50 per cent. Pulmonary or systemic infarcts from dislodged thrombi occur in one-third or more of the cases of auricular fibrillation as a terminal event. Most of these form in the left auricle, and particularly its appendix. Experimentally in animals, the minute-output of the heart decreases about 20 per cent with the onset of auricular fibrillation. In patients with mitral stenosis the restoration of normal rhythm increases the minute-volume output about 30 per cent.

Mitral stenosis constitutes about 25 per cent of all cases of valvular disease. Because of hemoptysis it was once thought to be related to pulmonary tuberculosis, but this has long ago been disproved. Certain other associations, however, appear to be true. The condition is occasionally associated with paroxysmal tachycardia. In such an event the patient may develop pulmonary congestion, cyanosis, and enlargement of the liver within a very short time after the paroxysm begins, but generally the tachycardia produces no permanent effect. Acute pulmonary congestion may occur, especially after exercise, and often is associated with hemoptysis. Death may result suddenly or be delayed for a few days. Rarely, anginal attacks occur, usually after extreme exertion and generally in the presence of aortic insufficiency.

It is highly probable that mitral stenosis, even in the most favorable cases, lessens slightly the lifespan of women who bear children. One woman may bear several children without apparent ill effect, and another die of congestive failure with the first or second child. This is probably proportional to the degree of mechanical handicap from the valve lesion or the myocardial damage.

Treatment: The essence of good treatment is wisdom and common sense. The discovery of the lesion does not warrant frightening the patient's family, or adding unnecessary restrictions to the patient. Reasonable exercise, especially in children, may be permitted, dependent on the degree of the circulatory handicap; but adults probably live longer if their occupations are sedentary. In extreme cases, patients probably should not marry. Moderation in the use of stimulants and tobacco, and the avoidance of late hours and overeating, are advisable. At the onset of auricular fibrillation or of congestive heart failure, the usual treatment for these conditions is needed. The chief problem is that of postponing congestive failure as long as possible. In patients with signs of congestive failure such as rapid pulse, dyspnea and fatigue, the administration of adequate doses of digitalis usually improves the cardiac function, even when the rhythm is regular.

Prognosis: With proper care, patients may be active even into their sixties. Those patients with "button-hole" mitral valve defects associated with other valve lesions and myocardial damage may die early; but even in uncomplicated cases the majority may not survive the fifth decade of life.

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